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IMAGES IN CARDIOLOGY

# Supracardiac Total Anomalous Pulmonary Venous Drainage With Giant Superior Vena Cava Aneurysm

## A Rare Combination

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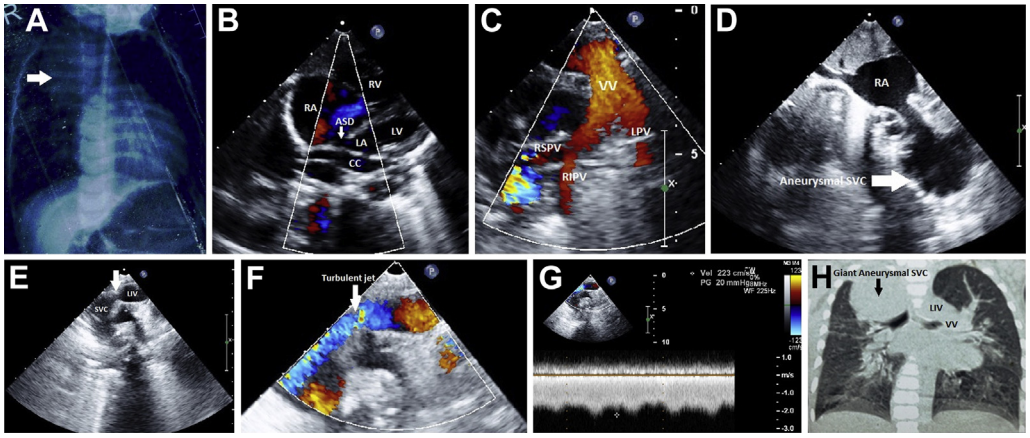


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A 9-month-old male child presented with cyanosis, recurrent respiratory infections, and failure to thrive. Oxygen saturation was 91%. There was a grade II/VI soft systolic murmur over the left upper sternal border. Chest radiography showed a prominent right upper mediastinal bulge (**A**, **arrowhead**) and mild cardiomegaly. Echocardiography revealed all pulmonary veins forming a common chamber (CC) behind the left atrium (LA) (**B**, Online Video 1), via the left vertical vein (VV) draining into the left innominate vein (LIV) **(C)**, and ultimately into the aneurysmally dilated superior vena cava (SVC) (**D**, Online Videos 2 and 3), suggestive of supracardiac total anomalous pulmonary venous connection (SCTAPVC). There was significant anatomical narrowing at the junction of the LIV and the SVC, with a pressure gradient (PG) of 20 mm Hg **(E and F)**. Contrast computed tomography demonstrated SCTAPVC with a giant aneurysmal SVC measuring 50.2 mm and stenosis at the junction of the SVC and the LIV **(G and H)**.

Aneurysmal dilation of the SVC is a rare anomaly, and its association with total anomalous pulmonary venous connection is rarer still. To the best of our knowledge, only 1 such case has been reported, with ours being the largest SVC aneurysm in association with SCTAPVC to date. In this case, dilation may have been secondary to the turbulent jet via the stenotic orifice to the wall of the SVC. This finding is important in planning surgical management at the time of SCTAPVC repair. ASD = atrial septal defect; LPV = left pulmonary vein; LV = left ventricle; RA = right atrium; RIPV = right inferior pulmonary vein; RSPV = right superior pulmonary vein; RV = right ventricle; vel = velocity.